In-Depth

Overview

Points To Remember About Scleroderma

- Scleroderma causes thick, hard patches of skin.
- Symptoms of scleroderma vary a lot, depending on the type of disease you have.
- There is no cure for scleroderma, but you can reduce damage from the disease with proper treatment.
- Since some types of scleroderma can cause serious damage to your internal organs, it is important to monitor your health and let your doctor know if you notice any new symptoms.

Scleroderma is the name of a group of diseases that cause your skin and sometimes other tissues to produce too much collagen, causing patches of tight, hard skin. Some forms of scleroderma can also damage your blood vessels and internal organs, such as the heart, lungs, and kidneys.

Derived from the Greek words “sklerosis,” meaning hardness, and “derma,” meaning skin, scleroderma literally means “hard skin.”

Scleroderma is called both a rheumatic (roo-MA-tik) disease and a connective tissue disease. The term rheumatic disease refers to a group of conditions characterized by inflammation or pain in the muscles, joints, or fibrous tissue. A connective tissue disease is one that affects tissues such as skin, tendons, and cartilage.

Scleroderma is also believed to be an autoimmune disease. In autoimmune diseases, the body's immune system turns against and damages its own tissues.

There is no cure for scleroderma, but treatment can improve your quality of life. People with diffuse scleroderma face the most serious long-term outlook if they develop severe kidney, lung, digestive, or heart problems. Fortunately, less than one-third of patients with diffuse disease

develop these severe problems. Early diagnosis and continual and careful monitoring are important.

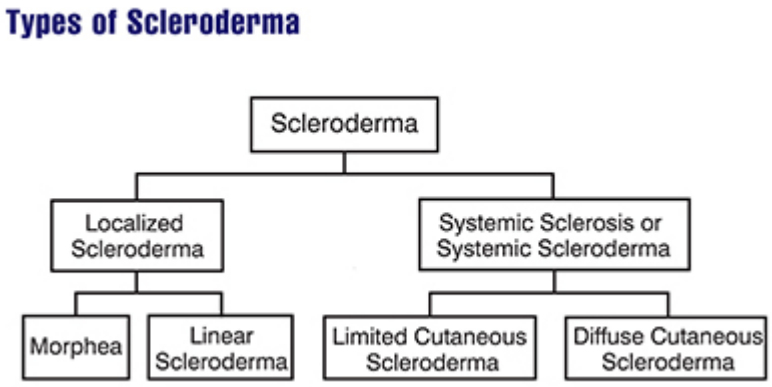
Who Gets

Anyone can get scleroderma, although it is more common in adults and women than in men and children. It affects people of all races and ethnic groups. However, there are some patterns by disease type. For example:

- Localized forms of scleroderma are more common in people of European descent than in African Americans. Morphea usually appears between the ages of 20 and 40, and linear scleroderma usually occurs in children or teenagers.
- Systemic scleroderma, whether limited or diffuse, typically occurs in people from 30 to 50 years old. It affects more women of African American than European descent.

Types

Some types of scleroderma are localized, meaning they only affect part of your body, and some are systemic, meaning they can affect your whole body. There are several different ways that doctors classify different types of scleroderma; here is one common way to look at it:



- Localized scleroderma, which affects the skin and related tissues, and generally appears in one or both of these patterns:
  - Morphea, or patches of scleroderma that may be a half-inch to a foot in diameter.
  - Linear scleroderma, or a line of scleroderma that usually runs down an arm or leg, but sometimes runs down the forehead.
- Systemic scleroderma, sometimes called systemic sclerosis, which affects your skin, tissues, blood vessels, and major organs. Doctors usually divide systemic scleroderma into two types:
  - Limited cutaneous scleroderma, which comes on gradually and affects the skin on your fingers, hands, face, lower arms, and legs.
  - Diffuse cutaneous scleroderma, which comes on suddenly and spreads quickly over

much of your body. This type can also cause organ damage.

Symptoms

The symptoms of scleroderma vary a lot depending on what type of scleroderma you have.

Localized scleroderma typically causes patches of thick, hard skin in one of two patterns.

- In one pattern, called morphea, you may have patches of reddish skin that thicken into firm, oval-shaped areas. These patches may stay in one area or spread to other parts of your body. The patches generally go away after three to five years, but you may still have darkened patches of skin and, in rare cases, weakened muscles.
- Another pattern of localized scleroderma is called linear scleroderma. This causes lines of thickened or different colored skin, usually down your arm or leg, but sometimes on your forehead.

Systemic scleroderma may come on quickly or gradually and may also cause problems with your organs and blood vessels.

- The kind that comes on gradually is called limited cutaneous scleroderma. It typically affects skin on your fingers, hands, face, lower arms, and legs. It can also cause problems with your blood vessels and esophagus.
- Diffuse cutaneous scleroderma comes on suddenly, usually with skin thickening on your hands. The skin thickening then spreads to the rest of your body. This is the type that can damage your internal organs, such as your intestines, lungs, heart, and kidneys. Fortunately, less than one-third of patients with diffuse cutaneous scleroderma have these serious problems.

Causes

Although scientists don’t know exactly what causes scleroderma, they are certain that people cannot catch it from or transmit it to others. Scientists suspect that scleroderma comes from several factors that may include:

- Abnormal immune or inflammatory activity. Like many other rheumatic disorders, scleroderma is believed to be an autoimmune disease. An autoimmune disease is one in which the immune system, for unknown reasons, turns against your own body.
- Genetic makeup. Although genes seem to put certain people at risk for scleroderma and play a role in its course, the disease is not passed from parent to child like some genetic diseases.
- Environmental triggers. Researchers suspect that exposure to some environmental factors may trigger scleroderma.
- Hormones. Women develop most types of scleroderma more often than men. Scientists suspect that hormonal differences between women and men might play a part in the

disease.

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Diagnosis

To determine whether you have scleroderma, your doctor will probably ask about your medical history and perform a physical exam. Your doctor may also order lab tests to check for certain antibodies that are common in people with scleroderma or do a skin biopsy.

Diagnosing scleroderma is easiest when you have typical symptoms and rapid skin thickening. In other cases, a diagnosis may take months, or even years, as the disease unfolds and reveals itself and as your doctor is able to rule out some other potential causes of the symptoms.

People with limited cutaneous scleroderma often have all or some of the symptoms that some doctors call CREST, which stands for the following symptoms:

- Calcinosis (KAL-sin-OH-sis), which is the formation of calcium deposits in the connective tissues, which can be detected by x-ray.
- Raynaud’s (ray-NOHZ) phenomenon, a condition in which the small blood vessels of the hands or feet contract in response to cold or anxiety.
- Esophageal (eh-SOFF-uh-GEE-ul) dysfunction, which means the impaired function of the esophagus (the tube connecting the throat and the stomach) that occurs when smooth muscles in the esophagus lose normal movement.
- Sclerodactyly (SKLER-oh-DAK-till-ee), which is thick and tight skin on the fingers, resulting from deposits of excess collagen within skin layers.
- Telangiectasia (tel-AN-jee-ek-TAY-zee-uh), a condition caused by the swelling of tiny blood vessels, in which small red spots appear on the hands and face.

Treatment

Your treatment will depend on which tissues and organs your scleroderma is affecting. Currently, there is no treatment that controls or stops the underlying problem – the overproduction of collagen – in all forms of scleroderma, but treatment can relieve symptoms and limit damage.

Your doctor may recommend a combination of medications, diet and lifestyle changes, and physical therapy to treat pain, skin dryness, and digestion problems.

Here are some more serious complications that develop for some people with scleroderma, and some of the ways that doctors typically treat them:

Lung damage: Virtually all people with systemic sclerosis have some loss of lung function. Some develop severe lung disease, which comes in two forms: pulmonary fibrosis (hardening or scarring of lung tissue because of excess collagen) and pulmonary hypertension (high blood pressure in the artery that carries blood from the heart to the lungs). Treatment for the two conditions is different:

- Pulmonary fibrosis may be treated with drugs that suppress the immune system, along with low doses of corticosteroids.
- Pulmonary hypertension may be treated with drugs that dilate the blood vessels or with newer medications that are prescribed specifically for treating pulmonary hypertension.

Regardless of your particular lung problem or its medical treatment, your role in the treatment process is essentially the same. To minimize lung complications, work closely with your medical team. Do the following:

- Watch for signs of lung disease, including fatigue, shortness of breath or difficulty breathing, and swollen feet. Report these symptoms to your doctor.
- Have your lungs closely checked, using standard lung-function tests, during the early stages of skin thickening. These tests, which can find problems at the earliest and most treatable stages, are needed because lung damage can occur even before you notice any symptoms.
- Get regular flu and pneumonia vaccines as recommended by your doctor. Contracting either illness could be dangerous for a person with lung disease.

Heart problems: Common among people with scleroderma, heart problems include scarring and weakening of the heart (cardiomyopathy), inflamed heart muscle (myocarditis), and abnormal heartbeat (arrhythmia). All of these problems can be treated. Treatment ranges from drugs to surgery and varies depending on the nature of the condition.

Kidney problems: Renal crisis is an uncommon but serious complication in patients with systemic sclerosis. Renal crisis results in severe uncontrolled high blood pressure, which can quickly lead to kidney failure. It’s very important that you take measures to identify and treat the hypertension as soon as it occurs. These are things you can do:

- Check your blood pressure regularly. You should also check it if you have any new or different symptoms such as a headache or shortness of breath. If your blood pressure is higher than usual, call your doctor right away.

- If you have kidney problems, take your prescribed medications faithfully. In the past two decades, drugs known as ACE (angiotensin-converting enzyme) inhibitors have made scleroderma-related kidney failure a less threatening problem than it used to be. But for these drugs to work, you must take them as soon as the hypertension is present.

Who Treats

Because scleroderma can affect many different organs and organ systems, you may have several different doctors involved in your care. Typically, care will be managed by a rheumatologist (a doctor specializing in treatment of musculoskeletal disorders and rheumatic diseases). Your rheumatologist may refer you to other health care providers, depending on the specific problems you are having. These specialists may include:

- Dermatologists, who treat skin problems.
- Orthopaedists, who specialize in the treatment of, and surgery for, bone and joint diseases or injuries.
- Pulmonologists, who treat lung problems.
- Nephrologists, who treat kidney problems.
- Cardiologists, who treat heart problems.
- Gastroenterologists, who treat digestive problems.

Typically, a rheumatologist will manage your care and refer you to specialists to treat specific areas of your body affected by the scleroderma. In addition to the doctors listed above, professionals such as nurse practitioners, physician assistants, physical or occupational therapists, psychologists, and social workers may play a role in your care. Dentists, orthodontists, and speech therapists can treat oral complications that arise from thickening of tissues in and around your mouth and on your face.

Living With

You can take an active part in treating your scleroderma. In addition to keeping appointments with your doctors and physical therapists, some of the suggestions below may help alleviate some of your symptoms.

Skin problems from scleroderma can make your skin dry and stiff. If your skin is affected, some of these ideas may help:

- Apply oil-based creams and lotions frequently, and always right after bathing.
- Apply sunscreen before you venture outdoors to protect against further damage from the sun’s rays.
- Use humidifiers to moisten the air in your home in colder winter climates. Clean humidifiers often to stop bacteria from growing in the water.
- Avoid very hot baths and showers, as hot water dries the skin.

- Avoid harsh soaps, household cleaners and caustic chemicals, if at all possible. Otherwise, be sure to wear rubber gloves when you use such products.
- Exercise regularly. Exercise, especially swimming, stimulates blood circulation to affected areas.

Scleroderma can make your mouth dry and damage connective tissues in your mouth, speeding up tooth decay and causing your teeth to become loose. Tightening facial skin can also make your mouth opening smaller and narrower, which makes it harder to care for your teeth. Here are some ways to avoid tooth and gum problems:

- Brush and floss your teeth regularly. If hand pain and stiffness make this difficult, consult your doctor or an occupational therapist about specially made toothbrush handles and devices to make flossing easier.
- Have regular dental checkups. Contact your dentist immediately if you experience mouth sores, mouth pain, or loose teeth.
- If decay is a problem, ask your dentist about fluoride rinses or prescription toothpastes that remineralize and harden tooth enamel.
- Consult a physical therapist about facial exercises to help keep your mouth and face more flexible.
- Keep your mouth moist by drinking plenty of water, sucking ice chips, using sugarless gum and hard candy, and avoiding mouthwashes with alcohol. If dry mouth still bothers you, ask your doctor about a saliva substitute or medications that can stimulate the flow of saliva.

Systemic sclerosis can affect any part of the digestive system. You may experience problems such as heartburn, difficulty swallowing, feeling full when you've just started eating, or intestinal complaints such as diarrhea, constipation, and gas. If your intestines are damaged, your body may have difficulty absorbing nutrients from food. Here are some things that might help at least some of the problems you have:

- Eat small, frequent meals.
- To keep your stomach contents from backing up into your esophagus, stand or sit for at least an hour (preferably two or three hours) after eating.
- When it is time to sleep, keep the head of your bed raised using blocks.
- Avoid late-night meals, spicy or fatty foods, alcohol, and caffeine, which can aggravate GI distress.
- Eat moist, soft foods, and chew them well. If you have difficulty swallowing or if your body doesn't absorb nutrients properly, your doctor may prescribe a special diet.
- Ask your doctor about medications for problems such as diarrhea, constipation, and heartburn. Some drugs called proton pump inhibitors are highly effective against heartburn. Oral antibiotics may stop bacterial overgrowth in the bowel, which can be a cause of diarrhea in some people with systemic sclerosis.

Research Progress

Research is finding better ways to treat symptoms, prevent organ damage and improve the

quality of life for people with scleroderma. Multidisciplinary research has also provided new clues for understanding the disease, which is an important step toward prevention and cure.

Studies of the immune system, genetics, cell biology, and molecular biology have helped reveal the causes of scleroderma, improve existing treatment and create entirely new treatment approaches.

Here's what some recent studies have found:

- Building on research that identified a gene associated with scleroderma in Oklahoma Choctaw Native Americans, scientists are using new technology to look for other genes associated with the disease's development and severity.
- The drug cyclophosphamide has been found effective in treating lung fibrosis. One study suggested that treating lung problems early with this immunosuppressive drug may help prevent further damage and increase chances of survival. Clinical trials assessing the effectiveness of other medications for lung fibrosis are ongoing.
- ACE inhibitors are used increasingly for scleroderma-related kidney problems. ACE inhibitors have greatly reduced the risk of kidney failure in people with scleroderma.
- Several drugs are now available to treat pulmonary hypertension. Previously, pulmonary hypertension was associated with a poor outcome, but medications have increased the quality of life and life expectancy for people with this dangerous form of lung damage.

Here are some areas of ongoing research:

- Researchers are looking at the complex immune system trying to determine why the body creates antibodies against its own tissues. Identifying where and when the antibodies are created and how they function could lead researchers to find targets for new therapies. Researchers are already trying new therapies, including those that suppress specific parts of the immune system.
- Studies continue to find the genes that predispose a person to getting scleroderma. Those genes may be targets for future therapies. Researchers are also trying to find genes (and other indicators) that could tell doctors who will develop what type of scleroderma. This could someday help doctors identify who might benefit from specific therapies. Additionally, since genes are not the only factor involved in scleroderma, researchers are looking for environmental factors that might trigger the disease in those who are genetically predisposed.
- Some scientists are trying to make better drugs to treat scleroderma and its symptoms. Other researchers are testing existing drugs to see if they are successful in delaying cardiovascular changes.
- Experiments using mice have proved valuable in past research on scleroderma. Further research using existing strains of mice continues.
- Researchers are looking into the many factors that lead to fibrosis to see if they can block its development.
- Studies have shown that certain chemicals called cytokines, made from cells in the body, contribute to inflammation. Researchers are trying to determine how cytokines contribute to the disease process.

Related resources

U.S. Food and Drug Administration
Toll free: 888-INFO-FDA (888-463-6332)
Website: https://www.fda.gov

Drugs@FDA at https://www.accessdata.fda.gov/scripts/cder/daf. Drugs@FDA is a searchable catalog of FDA-approved drug products.

Centers for Disease Control and Prevention, National Center for Health Statistics
Website: https://www.cdc.gov/nchs

American Academy of Dermatology
Website: https://www.aad.org

American College of Rheumatology
Website: https://www.rheumatology.org

Scleroderma Foundation
Website: https://www.scleroderma.org

Scleroderma Research Foundation
Website: https://www.srfcure.org

Arthritis Foundation
Website: https://www.arthritis.org

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